MECHANICAL EXSUFFLATION, NONINVASIVE VENTILATION, AND NEW STRATEGIES FOR PULMONARY REHABILITATION AND SLEEP DISORDERED BREATHING

JOHN R. BACH, M.D.

Assistant Professor of Physical Medicine and Rehabilitation
New Jersey Medical School-UMDNJ
Newark, New Jersey

Sir Patrick: Don't misunderstand me, my boy, I'm not belittling your discovery. Most discoveries are made regularly every fifteen years; and it's fully a hundred and fifty since yours was made last. That's something to be proud of . . .

Bernard Shaw, *The Doctor's Dilemma* (1906)

PATIENTS WITH CHRONIC OBSTRUCTIVE pulmonary disease or paralytic restrictive pulmonary syndromes including those with neuromuscular disease, kyphoscoliosis or traumatic quadriplegia may require frequent hospitalization because of respiratory impairment. Once ventilatory assistance becomes necessary, many patients never return to the community. In patients with intrinsic pulmonary disease, respiratory insufficiency initially results from failure of the respiratory mechanism adequately to oxygenate the blood and therefore tissues. Hypoventilation with elevated carbon dioxide levels develops only in late stages or during acute periods of respiratory encumberment. In those with paralytic restrictive syndromes, however, hypoventilation results from what is primarily ventilatory impairment, and hypoxia develops only secondarily. Although these conditions are pathophysiologically distinct and require entirely different therapeutic approaches, to the detriment of their patients, physicians all too commonly treat them as though they were the same. Moreover, physiatric principles that can benefit patients by reducing the number and length of hospitalizations and reduce the cost of maintaining patients in the community are generally ignored.

FORCED MECHANICAL EXSUFFLATION

Greater than 5 liters per second of peak expiratory flow is required to generate effective expulsion of airway secretions. Individuals with severe

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Address for reprint requests: Department of Rehabilitation Medicine, University Hospital B-239, 150 Bergen Street, Newark, NJ 07103

paralytic restrictive pulmonary syndromes may have less than 1 liter per second of peak expiratory flow. This is because of weak respiratory muscles in general, but many patients with amyotrophic lateral sclerosis, Duchenne muscular dystrophy, myotonic dystrophy, and myasthenia gravis also have significantly greater weakness of expiratory than of inspiratory muscles.[1] Clearance of airway secretions is further compromised when oropharyngeal muscle weakness complicates weakness of inspiratory and expiratory muscles. Patients with neuromuscular disease are only initially treated for ventilatory insufficiency and impaired coughing mechanisms when they cannot be weaned from mechanical ventilation following an episode of pneumonia caused by inability to clear secretions. Likewise, many spinal cord injured patients have virtually intact oropharyngeal and inspiratory muscle capacity, but generate expiratory flows inadequate to clear airway secretions. For these patients, too, ineffective airway clearance mechanisms become life-threatening during otherwise minor upper respiratory tract infections. Indeed, largely because of this, pulmonary failure is the most frequent cause of death during at least 11 years following injury in this population.[2] Postabdominal surgery patients are also at risk of serious respiratory complications because of transient expiratory muscle impairment.

Airway suctioning via nose or mouth does not effectively mobilize deep secretions and is poorly tolerated. Although tracheostomies have been placed in many patients only for the aspiration of airway secretions, but are usually refused electively. Tracheal suctioning causes irritation, increases secretions, and may be accompanied by severe hypoxia[3] and possibly cardiac arrhythmia. The suction catheter usually will not enter the left primary bronchus and will, therefore, not help to clear the left bronchial tree.[4] Tracheal suctioning at best effectively clears only tracheal secretions. It is ineffective in clearing life-threatening mucus plugs that adhere between the tracheostomy tube and tracheal wall and cuff. Chest percussion and postural drainage are commonly prescribed but are in themselves not adequate to clear the bronchial tree. Various techniques of manual assisted coughing are effective but underutilized (Figure 1).[5] These techniques are enhanced by preceding manually assisted exsufflation by an often necessarily assisted deep insufflation. A positive pressure blower (Zephyr, Lifecare, Lafayette, CO), intermittent positive pressure breathing machine, or portable ventilator is useful in delivering maximum tolerated insufflations for this purpose.

We have found that a mechanical exsufflation device is more effective than manual assisted coughing or tracheal suctioning and considerably less effort intensive. An original mechanical exsufflation device, the Col-flator, was

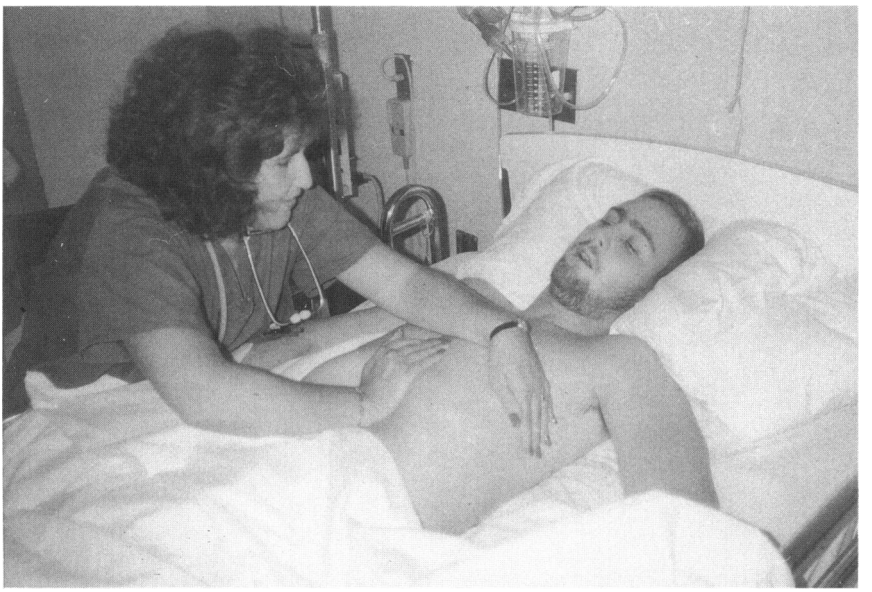

Fig. 1. Manual assisted coughing, most effective when preceded by a deep breath or mechanically assisted insufflation.

described in 1953 and then studied extensively.[6–9] This device delivers a deep, adjustable insufflation via an anesthesia mask or tracheostomy tube. Insufflation is followed by an independently adjustable exsufflation caused by a sudden drop in pressure of about 80 mm Hg in 0.02 seconds. The negative pressure is usually sustained for 1.5 to 2 seconds, and generates 5 to 11 liters per second of peak expiratory flow. This is more than can be generated by many physically intact individuals, and is thus optimal for clearing airway secretions.

Many ventilator-dependent patients unable to cough effectively continue to use this device to clear airway secretions during respiratory tract infections. We have had more than 50 nontracheostomized ventilator-assisted patients who have used the Cof-flator as necessary during respiratory infections without any complications for more than 30 years. It has also been used successfully by postabdominal surgery patients and patients with chronic obstructive pulmonary disease. The device has become an essential patient management tool in our intensive care units (Figure 2). It permits earlier extubation of intubated or tracheostomized patients during ventilator weaning, and can be instrumental in permitting continued ventilatory support by noninvasive ventilatory aids during intercurrent respiratory tract infections.

Although safe and very effective, manufacture of this device was discontin-

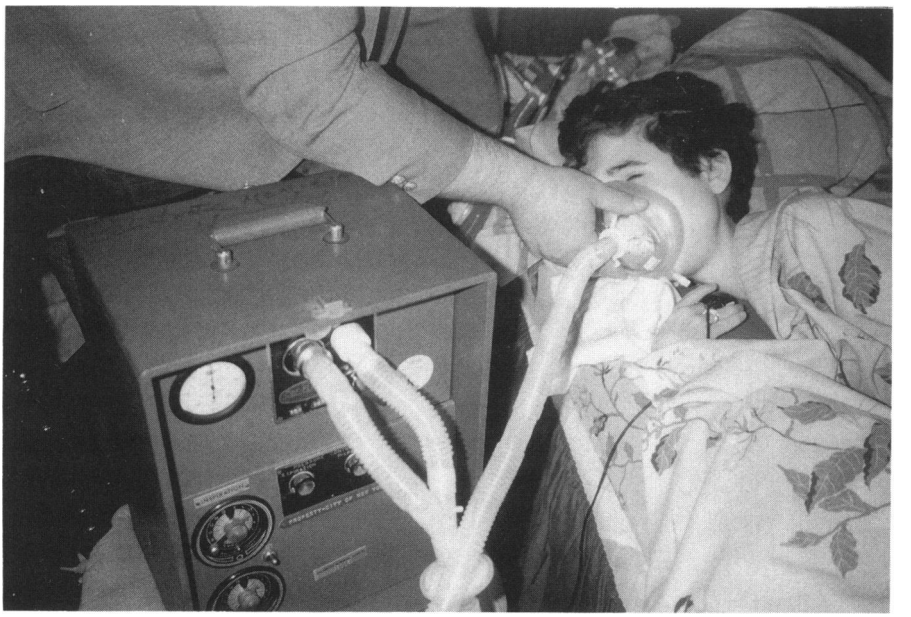

Fig. 2. A forced mechanical exsufflation device which delivers an adjustable deep insufflation followed by an adjustable sustained exsufflation to clear airway secretions.

ued in 1965 because of an inadequate market as a result of widespread resort to tracheostomy for suctioning. It was, nonetheless, available for rental until lack of replacement parts made this impractical in 1988. Its application should become vital, however, as intermittent positive pressure ventilation via nasal or oral access for nocturnal ventilatory support and oral access for daytime aid become more widely recognized.

A second form of forced mechanical exsufflation, the Jamil, is based on the principle of the intermittent abdominal pressure ventilator,[10] and is used predominantly by patients with cystic fibrosis and other obstructive pulmonary conditions in Europe.[11] The Jamil is an abdominal corset that encircles the abdomen and lower thorax. It is tugged by a motor driven unit with adjustable force and timing thereby assisting ventilation as well as providing forced exsufflation.

Pulse Oximetry Biofeedback and Respiratory Tract Infections

Pulse oximetry biofeedback and mouth intermittent positive pressure ventilation have been useful tools in weaning patients from ventilatory support and maintaining them free of hospitalization for respiratory impairment. Mouth intermittent positive pressure ventilation is an excellent alternative to

pressure support ventilation or interspersing periods of tracheostomy intermittent positive pressure ventilation with continuous positive airway pressure for weaning patients.[12,13] During daytime training periods a pulse oximeter is used and the oxyhemoglobin saturation (SaO_2) alarm usually set at 94% to 95%. The patient is instructed to maintain his SaO_2 at or above this level all day. He can either achieve this by unassisted breathing, or once he tires, by mouth intermittent positive pressure ventilation, usually by a portable ventilator. The patient sees immediately that by taking slightly deeper breaths his SaO_2 will exceed 95% within seconds. Thus he can be safely extubated or have his fenestrated tracheostomy tube plugged.

For a patient with chronic alveolar hypoventilation who has not been using ventilatory support, introduction to and use of mouth intermittent positive pressure ventilation with oximetry biofeedback guidance can facilitate resetting of respiratory control centers. This is further advanced by providing mouth intermittent positive pressure ventilation during sleep by use of a Bennett lip seal and determining its efficacy by monitoring SaO_2.[12–15]

The 24 hour attention of well-trained attendants or family members and use of mechanical exsufflation up to every 15 minutes as necessary permits even patients with little or no vital capacity to be safely managed at home during upper respiratory infections. Continuous SaO_2 monitoring is useful to evaluate or to assess the severity of atelectasis. This is seen as a continuous decrease in the SaO_2 baseline despite continuous ventilatory support by mouth or nasal intermittent positive pressure ventilation, or an effective body ventilator. Sudden mucus plugging, on the other hand, often causes an acute, sometimes severe, decrease in SaO_2. Once the mucus plug is cleared by mechanical exsufflation or other assist technique, the SaO_2 immediately returns to baseline. Many long-term ventilator dependent patients have become experts at training their attendants how and when to apply these measures.

Because of exacerbation of respiratory muscle weakness during respiratory infections,[16] the daily regimen of assisted ventilation invariably needs to be extended if it is not already 24 hours. Frequent "sighs" may be necessary for manual assisted coughing. Some patients on mouth or nasal intermittent positive pressure ventilation switch to a body ventilator for greater rest and easier access to mouth and nose to clear secretions. If hospitalization is to be avoided during respiratory infections, supplemental oxygen administration and sedatives should be avoided and broad spectrum antibiotics, adequate humidification, and hydration provided as necessary. On-call respiratory therapists of conscientious home care companies can play key roles in

maintaining ventilatory equipment, an exsufflation device, and an oximeter in the home, and in training the patient and his family how to use them.

Noninvasive Ventilatory Support

Acute life-threatening decompensations can be minimized and life significantly prolonged by noninvasive supported ventilation. Any patient with less than 50% of predicted normal supine vital capacity should undergo SaO_2 monitoring and noninvasive pCO_2 monitoring during sleep. The capnograph, which can be used to measure end-tidal pCO_2, and pulse oximeter must be capable of summarizing and printing out the data.[17,18] These studies are most conveniently performed on an ambulatory basis. Many patients with supine vital capacities less than 30% of predicted require at least nocturnal ventilatory support.[17] Patients with less than 12% of predicted vital capacity often require aid around the clock.[17] Since patients with chronic alveolar hypoventilation almost invariably refuse elective tracheostomy for intermittent positive pressure ventilation, acute ventilatory failure can be avoided only by noninvasive aids. Indications for their use include any cooperative intubated or tracheostomized patient with ventilatory insufficiency or any patient with chronic alveolar hypoventilation and the following: sufficient oropharyngeal muscle strength for an artificial inspiratory capacity[19] of at least 500 ml or adequate for swallowing, speaking, and clearing of oral secretions; no history of substance abuse; intact lung parenchyma with no need of supplemental oxygen to maintain a pO_2 greater than 60 mm Hg in the presence of normal pCO_2; access to effective means to clear airway secretions when necessary, i.e., a mechanical forced exsufflation device or an attendant trained in manual assisted coughing, no seizure disorder, and no orthopedic conditions that interfere with the use of a patient-ventilator circuit interface.

Ventilatory assistance or support can be provided by body ventilators, devices that act directly on the body, or intermittent positive pressure ventilation via mouth, nose, or oral-nasal interface. Negative pressure body ventilators create negative pressure on the chest and abdomen, and air then flows into the lungs through the nose and mouth. These devices include the Rocking Bed,[20] Iron Lung,[21] Porta Lung, wrap ventilator, and Chest Shell.[22–24] They are not feasible or are ineffective for use in the sitting position. Except for the Iron Lung and Porta Lung, they are generally not useful in the presence of scoliosis or extreme obesity. It may take 10 minutes or more for a personal care attendant to place a patient in a wrap ventilator. Sleeping intimately with a significant other is not possible. Travel with body ventilators is at best inconvenient, and these devices are associated with obstructive sleep apneas

in most patients using them.[25] For these reasons noninvasive intermittent positive airway pressure techniques, including most conveniently mouth and nasal intermittent positive pressure ventilation, are the methods of choice for long-term ventilatory support for most patients. Negative pressure body ventilators continue to be useful, however, for temporary assistance for some patients during acute respiratory infections,[14,15] during tracheostomy site closure,[12,13] and for patients who prefer them to noninvasive intermittent positive airway pressure techniques.

The intermittent abdominal pressure ventilator-consists of an inflatable bladder in an abdominal belt. The bladder is cyclically inflated by a positive pressure ventilator that pushes the abdominal contents up against the diaphragm and ventilates the patient. This generally augments the patient's tidal volume by 200 to 400 ml, but much greater volumes are often possible.[10] It is not effective in the presence of scoliosis, extremes of body weight,or when used in the supine position. It is most effective in the sitting position at 75–85°. It is the daytime ventilatory support preferred by most patients with less than one hour of free time from ventilatory assistance because it is cosmetic, practical, effective, and ideal for concurrent glossopharyngeal breathing and wheelchair use.[10]

Noninvasive intermittent positive airway pressure techniques are effective alternatives to tracheostomy intermittent positive pressure ventilation and body ventilator use. Mouth intermittent positive pressure ventilation is the method of daytime support preferred by most patients capable of one hour or more of free time, and is also preferred by patients with less than one hour of free time for whom the intermittent abdominal pressure ventilator is not effective. For daytime aid air is delivered to the patient through a mouth piece held in or adjacent to the mouth (Figure 3).[10,13–15] During sleep a Bennett lip seal is recommended for fixing the mouth-piece firmly in the mouth and minimizing insufflation leak from the mouth (Figure 4). Nocturnal administration of nasal intermittent positive pressure ventilation is preferred by most patients who require aid only during sleep.[17] Custom molded interfaces are useful for patients for whom the commercially available generic interfaces (CPAP masks) are uncomfortable or inadequate to prevent insufflation leak (Figure 5).[26] Oral-nasal interfaces are occasionally indicated, particularly for patients with difficulty donning or using the strap retention systems of mouth or nasal intermittent positive pressure ventilation.[18,26] Individually molded nasal interfaces, including the commercially available SEFAM mask (Lifecare, Lafayette, CO), may also be useful in management of obstructive sleep apnea.

Fig. 3. Mouth intermittent positive pressure ventilation, preferred by most ventilator dependent individuals who require daytime aid.

Versatility in the use of noninvasive ventilatory aids and effective management of intercurrent respiratory infections are paramount in averting unnecessary hospitalizations, delaying or avoiding tracheostomy and enhancing patient comfort. Averting difficulties in discharging ventilator-assisted individuals are illustrated in the following two cases. A 31-year-old woman with a history of poliomyelitis when four years old developed a respiratory infection and acute respiratory failure in April 1987.She had a cardiopulmonary arrest while en route to a local hospital, was intubated, then quickly had a tracheostomy. Attempts at ventilator weaning failed, and she was reluctantly transferred to a chronic nursing care facility although she required only nocturnal ventilatory support and had a vital capacity in the sitting position of 1,030 ml but only 630 ml supine. She was not permitted to return home to her husband and family because state law prohibited home management of tracheostomized ventilator-assisted individuals by other than skilled nurses, for whom no funds were available. She remained at the chronic care facility where she continued nocturnal tracheostomy intermittent positive pressure ventilation with an inflated cuff for six months. This made verbal communication impossible and caused severe tracheomalacia.

Upon transfer to a rehabilitation facility for removal of the tracheostomy

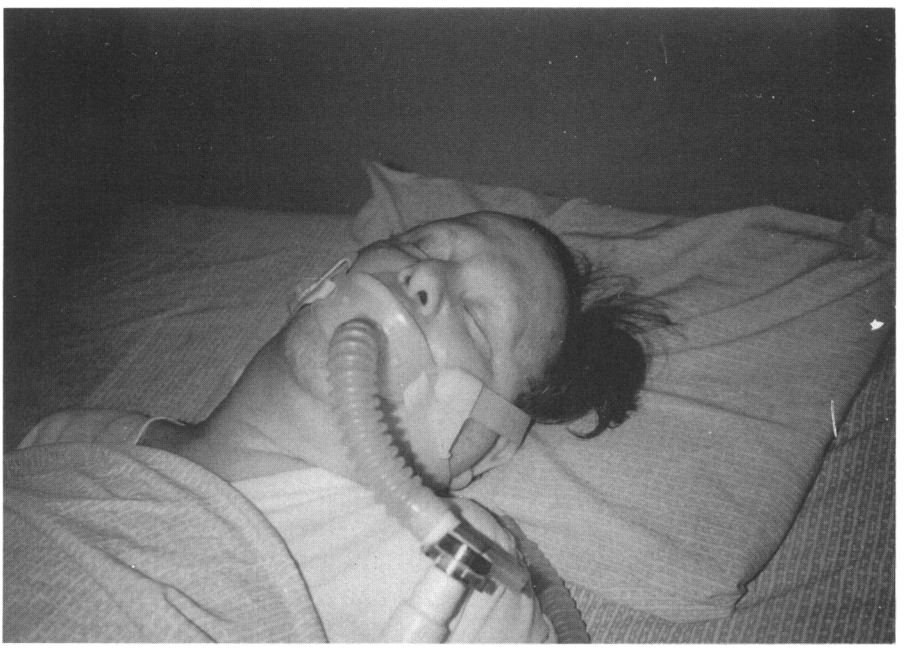

Fig. 4. Mouth intermittent positive pressure ventilation used during sleep with a Bennett lip seal.

tube, the first night her cuff was deflated and ventilator volumes increased to compensate for the insufflation leak across the vocal cords. This relieved pressure on the trachea and permitted speech. Later that week her tracheostomy tube was removed, a firm occlusive dressing placed over the site, and she was given nocturnal nasal intermittent positive pressure ventilation. She used this for two weeks until she complained of discomfort from the nasal interface. Once again an attempt at weaning failed when after 10 days off aid, severe fatigue and blood gas deterioration resulted in her being placed in an Iron Lung overnight for two weeks. She was converted to using a Pulmowrap Ventilator overnight and discharged home. Although this successfully ventilated her, it was inconvenient and she switched to a strapless oral nasal interface for ventilation. She continues to use this at night and has not required rehospitalization.

A 49-year-old woman with a congenital myopathy and a long history of fatigue, difficulties with concentration, dyspnea, and hypersomnolence developed pneumonia and was admitted to a New York City hospital and had a tracheostomy in February 1990. Weaned from intermittent positive pressure ventilation with the tracheostomy tube left open, she was given continuous supplemental oxygen therapy for three weeks but developed increasing hy-

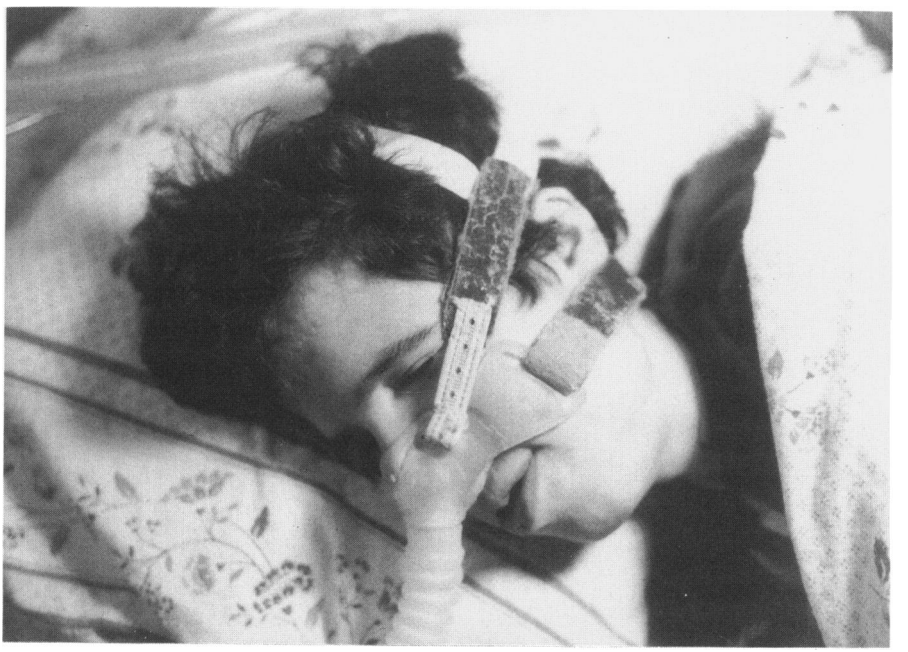

Fig. 5. An acrylic custom molded nasal interface for nocturnal support by nasal intermittent positive pressure ventilation.

percapnia (pCO_2 55 to 93 mm Hg). She could not tolerate plugging of the tube. Severe symptoms of hypoventilation lead to her return to tracheostomy ventilation. She continued to receive continuous oxygen therapy. Further attempts at ventilator weaning involved alternating the use of diminishing rate and periods of synchronized intermittent mandatory ventilation with continuous positive airway pressure. Her arterial blood gases typically demonstrated normal pH, pO_2 130 to 170 mm Hg, pCO_2 65 to 74 mm Hg and elevated bicarbonate levels.

After four months of failed weaning attempts, she was transferred to our service for tracheostomy removal July 27, 1990. Supplemental oxygen therapy, bronchodilators, and theophylline were permanently discontinued with no untoward effects. Oximetry biofeedback demonstrated that she could increase her SaO_2 to more than 95% and reduce her arterial blood gas pCO_2 to approximately 40 mm Hg with some extra effort. The tracheostomy tube was removed and she was ventilated by the Chest Shell Ventilator for 18 hours while the tracheostomy site closed sufficiently to permit the higher pressures of mouth or nasal intermittent positive pressure ventilation. Overnight while in the Chest Shell her mean SaO_2 was 89% with a low of 80% and the SaO_2 was below 90% 56% of the time, and below 85% 10% of the

time with a maximum recorded end-tidal pCO_2 of 55 mm Hg. She was uncomfortable and vomited twice while using the Chest Shell. She continued to maintain her SaO_2 at 95% or greater during the day by unassisted breathing supplemented by periods of mouth intermittent positive pressure ventilation. She was then given nasal intermittent positive pressure ventilation for nocturnal aid, during which time her mean SaO_2 was 97% with a low of 89%. Airway secretions due to tracheostomy tube irritation were cleared for the two days following tracheostomy site closure using a mechanical exsufflation device. She was discharged home the third day after admission on the regimen of mouth and nasal intermittent positive pressure ventilation just described. Although at discharge her maximum time free of the ventilator without dyspnea was less than two hours, maintenance of more normal ventilation around the clock kept her symptom free. She returned to her very active career as a professional writer and counselor.

Sleep Disordered Breathing

Sleep disordered breathing is a common entity that can develop into or complicate chronic alveolar hypoventilation, and that may also complicate chronic obstructive pulmonary disease. Sleep disordered breathing refers to the occurrence of apneas and hypopneas that may be centrally derived or result from upper airway obstruction. The obstructive sleep apnea syndrome in which apneas and hypopneas are primarily obstructive has potentially serious cardiovascular and neuropsychiatric sequelae.[27] Overt obstructive sleep apnea occurs in at least 3% of the general population, but its incidence increases greatly with age, in males, and with other endocrine, space occupying lesions, and neuromuscular conditions.[28] It also occurs in most patients with ventilatory insufficiency using negative pressure body ventilators[25,29] or electrophrenic nerve pacing.

Significant weight reduction can improve or completely resolve the obstructive sleep apnea syndrome in most obese patients.[29] However, this is all too frequently only temporarily accomplished.

Continuous positive airway pressure is effective for most patients. Independently varying inspiratory and expiratory pressures with the newly available BiPAP machine (Respironics Inc., Monroeville PA) improves effectiveness and comfort. Continuous positive airway pressure and variable inspiratory expiratory positive airway pressure may be ineffective, however, for many obesity hypoventilation patients or patients with a combined paralytic restrictive ventilatory insufficiency and concurrent obstructive sleep apnea. Assisted ventilation by nasal or mouth intermittent positive pressure venti-

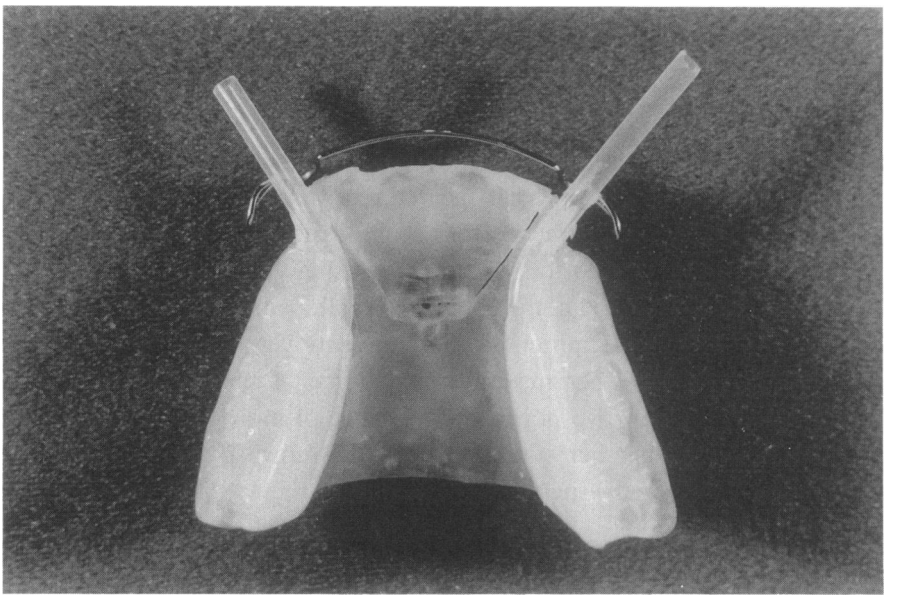

Fig. 6. An intra-oral appliance used to treat obstructive sleep apnea. Courtesy of Dr. John R. Haze, D.D.S., Montville, N.J.

lation as described earlier can both assist ventilation and maintain upper airway patency.

Continuous positive airway pressure, variable inspiratory expiratory positive airway pressure, as with previously described nasal intermittent positive pressure ventilation, can be delivered using commercially available CPAP masks (Respironics Inc., Monroeville PA; Puritan-Bennett Inc., Boulder CO). These inexpensive masks are often uncomfortable, and the key reason for the failure of patient compliance with long-term continuous positive airway pressure therapy in 43 of 125 recently studied patients.[30] Custom molded nasal interfaces, previously described, should then be considered.

A convenient long-term solution, effective for many obstructive sleep apnea syndrome patients, is an orthodontic splint that brings the mandible and tongue forward (Figure 6).[31,32] This can be effective and is commonly preferred. Supine positioning of the patient during sleep may also be effective.

Nasopharyngeal tubes and surgical options including tracheostomy, uvulopalatopharyngoplasty, and mandibular advancement procedures should be only a last resort.[33–35]

Chronic Obstructive Pulmonary Disease

The chronic obstructive pulmonary diseases include chronic bronchitis, emphysema, asthma, and cystic fibrosis, conditions that usually have signifi-

cant elements of both airway obstruction and parenchymal lung disease. Chronic obstructive pulmonary disease is the fifth leading cause of death in the United States. It affects 10–40% of all Americans and its incidence has doubled since 1970. Fifty percent of patients have activity limitations and 25% are bed disabled.[36] It is the fourth largest cause of major activity limitation. With the aging of the general population, a significant increase in its prevalence is projected in years to come.

Forced expiratory volume in one second (FEV_1) may decrease by 60 ml per year in chronic obstructive pulmonary disease patients, a rate almost twice normal. Exertional dyspnea occurs most often when the FEV_1 is less than 1,500 ml. Dyspnea and exacerbations of respiratory insufficiency associated with respiratory infections or other acute medical conditions are the most frequent reasons these patients are admitted to a hospital. This may occur several times during the course of a year. This is even more likely for patients who retain CO_2, and who thus have chronic alveolar hypoventilation as well as impaired oxygenation.

Reduction in dyspnea, increased maximum oxygen consumption and exercise tolerance, and fewer hospitalizations from respiratory impairment were observed in eight recently reviewed comprehensive pulmonary rehabilitation programs.[37] These significant improvements in quality of life may be sustained by continued rehabilitation goal-oriented activities. This is particularly important because patients with mild to moderate emphysema and chronic bronchitis without a major reversible component may survive 20 to 40 years.

A thorough presentation of the principles of evaluation and rehabilitation of chronic obstructive pulmonary disease patients is beyond the scope of this work. The reader is referred to standard text books devoted to this topic.[38–40] Three recent developments bearing on ambulatory management of these patients will be discussed: transtracheal oxygen delivery, respiratory muscle rest, and therapeutic exercise.

Transtracheal Delivery of Oxygen

Arterial pO_2 levels fall before an observed rise in pCO_2 because CO_2 diffuses more rapidly across the respiratory membrane. Thus, hypoxia in the presence of normal or hyperventilation (pCO_2 less than 43 mm Hg) characterizes intrinsic lung disease. Such patients may develop overt respiratory failure with or without hypercapnia during acute pulmonary infections. Oxygen therapy should be used for these patients as much as 24 hours a day if the pO_2 is less than 60 mm Hg. It decreases reactive pulmonary hypertension

and polycythemia, improves cognitive function, and may decrease the frequency of hospitalizations and prolong life. Transtracheal oxygen delivery is best for continuous use. It avoids waste around the nose and mouth, avoids the "dead space" of the nasopharynx, and prevents discomfort and drying associated with nasal cannulas and face masks.[38]

Exercise and Ventilatory Muscle Rest

Despite high ventilation rates in chronic obstructive pulmonary disease, ventilatory response to both hypercapnia and hypoxia may be reduced. This is often exacerbated during sleep. The increase in pulmonary vascular resistance that occurs in the presence of pulmonary tissue hypoxia is exacerbated by acidosis, and, when severe, leads to right ventricular failure. The use of oxygen therapy alone exacerbates CO_2 retention and acidosis. Such patients may have respiratory muscle fatigue and hypoventilation at least in part corrected by periods of respiratory muscle rest by assisted ventilation. Techniques to assist their ventilation are the same as those used by patients with paralytic restrictive pulmonary syndromes.

Braun and Marino[41] studied 14 patients with severe chronic airflow limitation and CO_2 retention treated at home by daily negative pressure assisted ventilation for at least five months. During periods of unassisted breathing these patients demonstrated a decrease in mean $PaCO_2$ from 54 mm Hg to 45 mm Hg, improvement in daytime arterial blood gases, improvement in vital capacity, relief of dyspnea, improved inspiratory and expiratory muscle strength, increased daily activity, and a seven-fold reduction in yearly hospitalizations. Others have shown that body ventilator use three to six hours per day, one to three days per week, can significantly improve pulmonary function and arterial blood gases.[42,43] However, yet to be published data on the nocturnal use of body ventilators appears to demonstrate no benefit. Since the majority of patients who use body ventilators overnight have significant periods of airway obstruction during sleep,[25] and since such obstruction will increase respiratory muscle effort rather than decrease it, it is likely that any benefit to be derived from body ventilator use will have to be derived from daytime use with the patient awake.

We have found that nasal and mouth intermittent positive pressure ventilation can provide respiratory muscle rest for many cooperative chronic obstructive pulmonary disease patients. Electromyographic monitoring of diaphragm activity during mouth or nasal ventilation demonstrates at least as much decrease in activity by these techniques as when using body ventilators. Since these methods are more convenient they may be preferred to the

use of body ventilators in the future. The patients with more severe CO_2 retention may benefit the most since CO_2 retention itself has been demonstrated to decrease muscle strength.[44]

Several studies have suggested better prognosis for chronic obstructive pulmonary disease patients since 1975 when long-term home tracheostomy intermittent positive pressure ventilation became more widespread.[45–48] As with patients with paralytic restrictive respiratory insufficiency, two groups of patients may be suited to ventilatory support at home. The first and smaller group includes those who require aid around the clock, usually by tracheostomy, but who are medically and psychologically stable. These patients tend, however, to require frequent hospital readmission and to have a poorer prognosis than ventilator dependent patients with neuromuscular disease.[49] The second group may benefit from nocturnal assistance alone. As with patients with progressive neuromuscular disease, patients with chronic hypoventilation (pCO_2 greater than 47 mm Hg) during sleep are candidates for nocturnal assisted ventilation. Although in patients with paralytic restrictive respiratory insufficiency, nocturnal ventilatory support alone can prevent cor pulmonale and episodes of acute respiratory failure,[14,15,17,18] most patients with chronic obstructive pulmonary disease require supplemental oxygen therapy as well.

For any kind of ventilatory assistance air trapping can be a limiting factor for chronic obstructive pulmonary disease patients. Perhaps the intermittent abdominal pressure ventilator and the previously described Jamil, which work by assisting exsufflation, have theoretical advantages over other forms of assisted ventilation when used for this patient population. These techniques have been demonstrated as useful in ventilating patients with severe intrinsic lung disease.[50] Unfortunately, they are only practical for long-term use by nonambulatory individuals. Further clinical study is warranted.

Although it is commonly observed that patients with mild to moderate disease do best in therapeutic exercise programs, positive results have been reported for exercise conditioning of a group of advanced chronic obstructive pulmonary disease patients with CO_2 retention:[51] 117 of the 120 patients studied had FEV_1 less than 1 liter. Following rehabilitation and exercise reconditioning, there were statistically significant 7 to 8% improvements in pulmonary function variables, including VC (120 ml), FEV_1 (40 ml), and maximum inspiratory pressures (3.1 cm H_2O). Arterial pCO_2, pO_2, pH, and maximum expiratory pressures did not change significantly. VC, FEV_1 and maximum inspiratory pressures did not vary as a function of initial pCO_2 levels but the higher the initial pCO_2 the more pCO_2 fell and pO_2 rose during

the rehabilitation program. Ambulation distance increased very significantly for all patients. Patients' ability to perform activities of daily living changed in parallel with the walking distance. That hypercapnic patients tolerated a rigorous reconditioning program suggests that exercise may not necessarily precipitate diaphragm fatigue in this population. Interestingly, this was the case despite the fact that hypercapnic patients with chronic obstructive pulmonary disease have weaker respiratory muscles than eucapnic patients.[52,53]

It may be that interspersing periods of respiratory muscle rest with exercise, a key principle in the pulmonary rehabilitation of patients with spinal cord injury,[54] may also be optimal for patients with chronic obstructive pulmonary disease. Mechanical assistive breathing devices should be used when pCO_2 exceeds 50 mm Hg or pO_2 fails to improve beyond 40 mm Hg despite inhalation of 100% oxygen. Improved pulmonary function and daytime gases, increased VC, decreased fatigue, and increased well-being have been reported in programs providing respiratory muscle rest with exercise reconditioning.[1,55]

Cost

In a recent survey of 453 ventilator-assisted individuals living in Illinois, 145 were in acute hospitals and 105 in extended-care facilities. Sixty percent of the patients had cardiopulmonary disorders. Forty percent had neuromuscular or musculoskeletal abnormalities, including spinal cord injuries. The monthly hospital charges averaged $22,190 with a range of $10,020 to $66,750. Most reimbursement was public. Barriers to discharging these patients to the community included inadequate payment for community-based services and lack of access to information.[56]

In New York City a Medicaid designated not-for-profit corporation permits self-directed disabled individuals to live in the community.[57] It aids its clients in finding and organizing home personal attendant care, and currently has 37 totally disabled ventilator supported clients of whom 30 depend on ventilatory support around the clock, but only seven have tracheostomies. The 30 patients using noninvasive aids depend on the intermittent abdominal pressure ventilator and mouth or nasal intermittent positive pressure ventilation with three additionally using body ventilators. The total cost, including all equipment, supplies, transportation, food, and attendant care for the home management of these individuals, amounts to $180 to $270 a day. This is less than one half the average cost for hospital care quoted by Goldberg,[56] and about 70% less than management in specially designated respiratory units of chronic care facilities in New York City. Further, the advantages to the

patients in terms of quality of life and facilitation of gainful employment are immeasurable. Patients with tracheostomies are not eligible for this program except with the presence of nursing care, which is not paid by public assistance programs, or an uncompensated family member for overnight care and tracheal suctioning.

SUMMARY

Manual and mechanical exsufflation are important but underutilized ways to clear airway secretions. These methods are especially useful when used in concert with noninvasive intermittent positive airway pressure ventilatory assistance to facilitate extubation and ventilator weaning. This can be used as much as 24 hours a day as an alternative to tracheostomy ventilation or body ventilator use for patients with paralytic restrictive ventilatory insufficiency. These techniques expedite community management of ventilator assisted individuals by avoiding tracheostomy and need for invasive suctioning and ongoing wound care. For these techniques to be effective and to prevent further suppression of ventilatory drive, supplemental oxygen administration must be avoided unless pO_2 is less than 60 mm Hg despite normalization of pCO_2. Custom molded interfaces for the delivery of noninvasive intermittent positive airway pressure ventilatory assistance can also be used to facilitate the delivery of variable inspiratory expiratory positive airway pressure for patients with obstructive sleep apnea. Noninvasive intermittent positive airway pressure ventilatory assistance or body ventilator use can rest the respiratory muscles of patients with advanced chronic obstructive pulmonary disease. This and pulmonary rehabilitation programs geared to exercise reconditioning are therapeutic options that significantly improve the quality of life of these patients. For both paralytic restrictive and obstructive pulmonary patients, these techniques decrease cost and frequency of hospitalizations.

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